

Brachial Plexus Lesion Due to Pathological Fracture of Clavicle in a Patient With Primary Hyperparathyroidism

Primer Hiperparatiroidizml Bir Hastada Klavikulanın Patolojik Kırığı Nedeniyle Meydana Gelen Brakiyal Pleksus Lezyonu

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Summary

Primary hyperparathyroidism (PHPT) is the third most common endocrine disorder. The clinical manifestations primarily involve the kidneys and the skeletal system. Pathologic bone fractures and proximal myopathies are rare conditions associated with PHPT. Early brachial plexus lesion secondary to clavicle fracture is very rare as well. The present case is of importance since it includes all of the three above-mentioned rare conditions. A 43-year-old female patient presented with complaints of pain and weakness in the right arm and limitation of movements in the right shoulder after a minor trauma. According to clinical, laboratory and radiological examination, she was diagnosed as having PHPT and traumatic right brachial plexus lesion due to pathologic fracture of clavicle, as well as hyperparathyroidism associated with secondary osteoporosis and myopathy. The patient underwent parathyroid surgery. After the surgery, a rehabilitation program including range of motion and strengthening exercises was given. PHPT may be diagnosed in an advanced stage of the disease. Therefore, clinicians should keep in mind that PHPT may present with complications of hyperparathyroidism.

Türk J Phys Med Rehab 2011;57 Suppl 2: 361-5.

Key Words: Primary hyperparathyroidism, pathologic fracture, brachial plexus lesion, myopathy

Özet

Primer hiperparatiroidizm (PHPT) üçüncü en sık görülen endokrin bozukluktur. Klinik bulgular, öncelikli olarak böbrekte ve iskelet sisteminde görülür. Patolojik kemik kırıkları ve proksimal miyopatiler PHPT ile ilişkili nadir görülen durumlardır. Klavikula kırığına ikincil olarak erken dönemde ortaya çıkan brakiyal pleksus lezyonu da çok nadirdir. Bu vaka yukarıda bahsedilen nadir görülen üç durumun tümünü içermesi nedeni ile önem taşımaktadır. Kırık üç yaşındaki kadın hasta minör travma sonrası sağ omuz hareketlerinde kısıtlılık, sağ kolda ağrı ve güçsüzlük şikayetleri ile başvurdu. Klinik, laboratuvar ve radyolojik bulgulara göre hasta PHPT ve klavikulanın patolojik kırığına bağlı travmatik sağ brakiyal pleksus lezyonu ve hiperparatiroidizm ile ilişkili ikincil osteoporoz ve miyopati olarak değerlendirildi. Hastaya paratiroidektomi operasyonu uygulandı. Paratiroidektomi ameliyatı sonrası, eklem hareket açıklığı ve güçlendirme egzersizlerini içeren rehabilitasyon programı verildi. PHPT tanısı hastalığın ileri evresinde konabilir. Klinisyenler PHPT'nin hiperparatiroidizmin komplikasyonları ile ortaya çıkabileceğini akıld tutmalıdırlar. *Türk Fiz Tıp Rehab Derg 2011;57 Özel Sayı 2: 361-5.*

Anahtar Kelimeler: Primer hiperparatiroidizm, patolojik kırık, brakiyal pleksus lezyonu, miyopati

Introduction

Primary hyperparathyroidism (PHPT) is the third most common endocrine disorder, with the highest incidence in postmenopausal women. Asymptomatic disease is common, severe disease with renal

stones and metabolic bone disease arises less frequently now than it used to be 20-30 years ago (1).

The clinical manifestations primarily involve the kidneys and the skeletal system. Skeletal involvement includes polyostotic lytic lesions and/or diffuse osteoporosis as a result of increased bone

resorption due to excessive production of parathyroid hormone (PTH). Osteitis fibrosa cystica (OFC) is an uncommon bone disease in PHPT and usually occurs in patients in the advanced stages of the disease. Low bone density and impaired bone quality predispose to pathologic fractures. Patients often require parathyroidectomy as a definitive treatment (2-4).

Patients with PHPT may have generalized mild muscle weakness. Clinically, hyperparathyroid myopathy includes proximal weakness and wasting, occasional bulbar weakness, and preserved or even brisk reflexes. The serum creatine phosphokinase (CPK) level is usually normal (5-7).

Herein we report a patient with advanced-stage PHPT who presented with proximal myopathy, pathologic clavicle and humerus fractures, as well as traumatic brachial plexus lesions secondary to clavicle fracture.

Case Report

A 43-year-old female patient was admitted to our outpatient clinic with complaints of pain and weakness in the right arm and limitation of movements in the right shoulder. It was noted from her history that she had sustained minor trauma to her right shoulder 4-5 months ago and pain and weakness had begun within a few days after the trauma. Since she has been living in a village, she had not consulted a physician right after the trauma. Because her symptoms did not resolve spontaneously, she had undergone a physical examination by a physician 2 months after the trauma. During this examination, she had a fracture in the right arm. The fracture was immobilized for 6 weeks.

The patient was admitted to our outpatient clinic because of her sustained complaints. There was no family history of any systemic disease, including endocrinopathies. In addition, there was no history of increased urinary frequency, urolithiasis, excessive thirst, chronic constipation, renal and peptic ulcer diseases, neck irradiation, and use of any medicines or vitamin supplements. On physical examination, a deformity involving the right humerus, atrophy in the right arm, and a little right scapular winging were noted (Figure 1). Range of motion of the right shoulder was limited. The muscle strength graded



Figure 1. Little scapular winging of the scapula which may be due to scapulo-thoracic dysfunction caused from inactivation of shoulder muscles.

according to the scale proposed by the British Medical Research Council (BMRC). It was 2/5 for right shoulder abductors, forearm flexors, whereas 3/5 for forearm extensors and 4/5 for shoulder elevators. Hypoesthesia was assessed on the lateral side of the right arm and the forearm, and the biceps and brachioradial reflexes were hypoactive. Although the muscle strength of the left upper extremity was markedly better than the right upper extremity, the strength of the proximal muscles of the left upper extremity was 4/5. The sensory and reflex examinations of the left arm were normal. The strength of the proximal muscles of the lower extremity was 4/5, and sensorial examination and reflexes were normal. Atrophy of the gluteal muscles and Trendelenburg gait were noted.

The blood pressure was 120/80 mmHg. While the levels of serum calcium (10.9 mg/dL; normal: 8.8-10.6 mg/dL), alkaline phosphatase (941 U/L; normal: 30-120 U/L), intact PTH (149 pml/mL; normal: 10-65 pml/mL), and osteocalcin (104 ng/mL; normal: 0.4-11 ng/mL) levels were high, the levels of phosphate (1.95 mg/dL; normal: 2.5-4.5 mg/dL) and 25-OH vitamin D (24 nmol/L; normal: 25-125 nmol/L) were low. Calcium and phosphorus levels in the 24-hours urine specimen were 242 mg/day (normal: 100-300 mg/day) and 23 mmol/day (normal: 12.9-42 mmol/day), respectively. The thyroid function tests and CPK levels were within normal limits. The hematologic and the remaining biochemical profiles including the protein electrophoresis and tumor markers were also normal. Urine Bence-Jones protein was negative. Urinalysis, chest radiography, and abdominal ultrasonography (US) were also normal.

On roentgenographic examination, a callus tissue of an old fracture in the middle 1/3 part of the right clavicle (Figure 2), and cystic lesions together with a non-union fracture were detected in the



Figure 2. A callus tissue of an old fracture in the middle 1/3 part of the right clavicle.

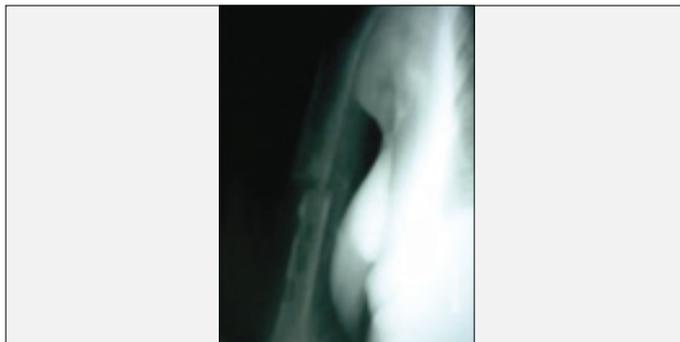


Figure 3. Cystic lesions and a non-union fracture in the middle part of the right humerus.

middle part of the right humerus (Figure 3). A brown tumor in the 4th metacarpal bone of the right hand with subperiosteal resorption and cystic lesions were observed (Figure 4). Dual energy X-ray absorptiometry (DXA) indicated diffuse severe osteoporosis with a L2-L4 mean T score of -5.0 and a femoral neck T score of -5.1.

On nerve conduction studies (NCS) of right arm, there was a mild reduction in sensory nerve conduction velocities in the right medial and the lateral antebrachial cutaneous nerves. Recording from the right deltoid and stimulating the axillary nerve at the Erb's point and recording from the right biceps brachii and stimulating the musculocutaneous nerve at the Erb's point produced a compound motor action potential with mildly prolonged latency and markedly decreased amplitude. Needle electromyography revealed increased numbers of long duration, high amplitude polyphasic motor unit potentials and discrete activity in the recruitment pattern of the right deltoid, biceps brachii, and the extensor digitorum communis muscles. To summarize, these findings included partial chronic axonal lesion of the upper and middle trunks of the right brachial plexus. The NCS of right long thoracic and dorsal scapular nerves as well as the right ulnar, median and radial nerves were within normal limits. Additionally, needle electromyography of the left deltoid, and the right and left gluteus medius muscles revealed short-duration polyphasic motor unit potentials with an increased recruitment pattern compatible with myopathy. Abnormal spontaneous activity was not observed in any of the evaluated muscles.

On US examination, an 11x33 mm nodular lesion was identified on the postero-inferior part of the right lobe of the thyroid gland. Scintigraphic findings compatible with a parathyroid adenoma on the median-inferior part of the right lobe were noted on technetium-99m-sestamibi-parathyroid scintigraphy.

In the light of these clinical and laboratory findings, the patient was diagnosed as having PHPT and traumatic right brachial plexus lesion due to pathologic fracture of clavicle, as well as hyperparathyroidism associated secondary osteoporosis and myopathy. She was transferred to the general surgery clinic and



Figure 4. Brown tumor in the 4th metacarpal bone of the right hand, subperiosteal resorption and cystic lesions.

underwent a subtotal thyroidectomy and parathyroid adenectomy operation. No postoperative complications have developed. The levels of calcium and alkaline phosphatase decreased to 9.3 mg/dL and 600 U/L, respectively, on the 4th postoperative day.

Histopathologic examination of the tissue sample removed during the operation was consistent with a parathyroid adenoma and Hashimoto thyroiditis. Due to the postoperative hypothyroidism, levothyroxine sodium (0.1 mg/day) was initiated with calcium and vitamin D. After surgery, the patient was taken to rehabilitation program consisting of range of motion exercises for the right shoulder and strengthening exercises for the proximal muscles. Marked improvement in the weakness of the proximal muscles was observed in the early postoperative period.

Although, monitoring with serial thyroid function tests, bone mineral density (BMD), and bone turnover markers were planned after discharge from the hospital, the patient discontinued her follow-up visits. On phone call, she expressed that she had no complaints.

Discussion

Primary hyperparathyroidism is a disorder of the parathyroid glands as a result of an excessive and inappropriate secretion of PTH, which was clearly recognized by von Recklinghausen in 1891 (8). The etiology of PHPT is predominantly chief cell adenomas. It is also infrequently related to diffuse hyperplasia or multiple adenomas; carcinoma is rare. The prevalence rate varies from 1/1000-4000, with a female:male ratio of 3:1 (1,4,8-11).

Elevated levels of serum calcium, alkaline phosphatase, and PTH, and decreased serum phosphate level are diagnostic indicators of PHPT, as in the present case (3,12).

The diagnosis of PHPT by routine automated biochemical screening has become commonplace in developed countries, thus, early diagnosis is possible in the disease course, even though it is often asymptomatic. On the other hand, in developing countries, PHPT is often seen in an advanced stage with bone involvement (4,13,14). Singhal et al. (15) reported that 5%-15% of patients with PHPT have associated bony abnormalities. The skeletal involvement includes diffuse osteoporosis or polyostotic lytic bone lesions, which are manifested clinically by diffuse or focal bone pain, or pathologic fractures involving osteolytic bone lesions (3,4,12). Radiologic examination reveals osteopenia of the entire skeleton and multiple localized lytic lesions with a benign aspect. Subperiosteal bone resorption of the phalanges is the most sensitive radiologic sign of PHPT (2,3,11).

Patients with PHPT have reduced BMD, especially at the cortical bone. The cortical width is constantly diminished and the cortical porosity is increased, whereas trabecular volume is normal and microarchitecture preserved (16-19). The DXA scan in our patient demonstrated osteoporosis. Because of the increase in BMD, which can last up to four years after parathyroidectomy operation, our patient should have been followed up for a long period (3,4,14,20).

Unfortunately, the patient described herein did not comply with the recommended follow-ups.

The classic skeletal manifestation of hyperparathyroidism is OFC (2). Brown tumors represent a severe form of OFC (21). The incidence of brown tumors in patients with PHPT is 1.5-1.7% (4,22). Brown tumors usually develop in the 3rd-4th decades of life, and females are more frequently affected. The brownish appearance results from hypervascularity, hemorrhage, and hemosiderin accumulation (23). Our patient had a brown tumor in the 4th metacarpal bone in the right hand.

Pathologic fractures secondary to PHPT may result from severe generalized osteopenia, or from localized bone destruction (4,24,25). A fracture out of proportion to the trauma, multiple fractures, as well as multiple sites of fracture in the same bone, are indicators of pathologic fractures (12,24). Our patient had two pathologic fractures in the right clavicle and humerus secondary to PHPT. In case of symptomatic PHPT and a pathological fracture, patients require parathyroidectomy while fracture treatment can be conservative, if undisplaced and uncomplicated (4).

Although the brachial plexus is vulnerable to trauma because of its superficial location, brachial plexus injury is a rare complication of fractured clavicle. When the trauma is severe, the fractured and displaced fragments may cause an acute severe lesion of the subclavicular neurovascular bundle. In limited lesions with minimal crush, as in the present case, it may appear clinically a few days after the trauma with a mild to moderate lesion. However, in the majority of the patients, neurologic symptoms develop later by large callus formation to encroach on the costoclavicular space (26-29). Depending on the location of injury, clinical presentations vary. In the present case as well, a pathologic fracture occurred in the middle 1/3 part of the clavicle after a minimal crush, and the lesions in the upper and medial trunks of brachial plexus were likely due to direct pressure of the fracture fragment. Early and direct compression of the brachial plexus by a fragment of clavicular bone is exceptional, and has rarely been reported (27).

Muscle weakness is a rare presenting feature of PHPT (7). Characteristically, weakness of proximal muscles, which tends to affect the pelvic girdle muscles earlier and more severely than the pectoral muscles, occurs. Distal muscle groups may also be affected in severe cases. The pathogenesis of hyperparathyroid myopathy remains undefined. Hypercalcemia reduces neuromuscular excitability and may cause muscle weakness. Serum muscle enzyme activities are usually normal, but creatinuria may be detected (5-7). In PHPT, muscle strength usually improves after surgery. In the present case, the proximal muscles were more affected in the lower extremities, whereas the distal muscle groups were normal. Short-duration polyphasic motor unit action potentials compatible with myopathy were determined on needle EMG performed on the proximal muscles of the lower and upper extremities. The serum CPK level was normal. Marked improvement in muscle strength was determined in the early postoperative period (5-7,30).

Parathyroid adenomas can be imaged by various radiological methods, including selective angiography, US, computed

tomography (CT), magnetic resonance imaging (MRI), and scintigraphy. CT and MRI are not routinely used in the localization of parathyroid adenomas due to high accuracy and wide availability of US and scintigraphy (2). In order to enhance the success of the surgery in the present case, location of the adenoma was determined using both US and technetium-99m-sestamibi-parathyroid scintigraphy techniques.

Although, pathologic fractures and brown tumors are extremely rare clinical phenomena in PHPT, in our case, both of them were seen. The formation of brachial plexus injury by direct pressure of the clavicular fracture on the brachial plexus has rarely been reported. In addition, muscle weakness is a rare presenting feature of hyperparathyroidism. The present case is of importance since it includes all of the three above mentioned rare conditions. Unfortunately, the present case was evaluated too late for optimal treatment. It is suggested that in a patient presenting with a pathologic fracture, routine measurements of serum calcium, phosphate, alkaline phosphatase, and intact PTH should be performed. This case report is also important in terms of indicating that a patient with PHPT may also present with complications due to pathologic fractures.

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